347 Fore arm bone characteristics in CF adolescents and young adults

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Introduction: In relation to longer survival, an increased rate of fractures has been seen in CF. We aimed to measure fore arm bone characteristics – related to bone fragility – by peripheral quantitative computed tomography (pQCT) and total body composition using dual-energy X-ray absorptiometry (DXA) to evaluate their degree of impairment and their potential interrelationships.
Methods: Body composition was measured by a Hologic 4500 DXA device. Cortical area, total BMD and thickness at the proximal site, as well as trabecular BMD at the distal site were measured by the same operator using a Stratec 2000 pQCT. Data are expressed as Z-scores using published references.
Results: Forty-eight patients (31 males and 17 females), between 12 and 30 years (mean 20), were studied. Main anthropometric features, including whole body fat mass and lean mass by DXA were within normal limits. BMD and cortical area z-scores were within normal references in males and females, whereas cortical thickness z-score of the proximal radius was significantly ($p < 0.05$) diminished [−1.22(0.84) vs −1.61(0.61)]. Whole body lean mass correlated more strongly with cortical thickness ($r = 0.72$, $p < 0.001$) than with total BMD ($r = 0.39$, $p < 0.05$).
Conclusions: Slight underweight adolescents and young adults with CF have preserved BMD but a reduced cortical bone size at the radius. Total body lean mass correlates better with fore-arm bone geometry parameters than BMD, suggesting that well-nourished CF patients may have reduced bone strength despite a normal BMD. Efforts to increase lean mass might also be helpful in the prevention of fractures in these patients.
Supported by: BVSM-ABLM.

348 CF osteopenia and bone tissue metabolism

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Introduction: Osteopenia is one frequent complication of CF.
Material and Methods: Close study of 40 patients at the age 9–24 of CF Center was made in order to investigate the rate of osteopenia and bone tissue metabolism. Method of dual energy X-ray absorptiometry ("Osteometer DTX-200", Denmark–USA) was used to measure BMD of forearm bone. Bone tissue metabolism was assessed by ELISA. A control group was represented by 35 healthy people of the same age group.
Results: BMD reduction was assessed in 62.5% of children and adults with CF, the rate of osteoporosis amounted to 15.6%. Compared to the control group the BMD rates are considerably lower in cases with CF (0.368 ± 0.018 and 0.515 ± 0.020 g/cm^2, $p < 0.001$). BMD rate of patients with osteoporosis is the lowest (0.321 ± 0.014 g/cm^2, $p < 0.001$). Patients with F508del genotype display Z-score rates (-2.75 ± 0.138 SD) and BMD (0.319 ± 0.007 g/cm^2) which are much lower than those of patients with "nonF508del" genotype (-0.73 ± 0.21 SD and 0.382 ± 0.02 g/cm^2, $p < 0.001$).
Bone tissue metabolism is characterized with suppressed process of bone remodeling which is indicated by the reduction of osteocalcin rate (52.73 ± 7.078 and 110.4 ± 9.3 ng/ml, $p < 0.001$) and C-end telopeptides (8.244 ± 1.292 and 17.34 ± 2.21 nmol/ml, $p < 0.001$) compared to the control group. Patients with F508del genotype suffer the growth of bone tissue metabolism impairment in adolescence due to disease severity progression.
Conclusions: Further research is seen in the direction of CF osteopenia correction with the complex of calcium and vitamin D3 and therapy of osteoporosis in adolescence and youth with osteogenon.

349 Influence of oral and inhaled corticosteroids treatment on bone mineral density (BMD) in patients with cystic fibrosis

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In our study we have analysed the influence of oral and inhaled corticosteroids on BMD in children with CF.
Aim: To reveal correlation between continuance of corticosteroids treatment and BMD.
Methods: 40 CF children (23F and 17M, 5–17.4 years) were included. All patients have "severe" phenotype. 14 children inhaled corticosteroids for 34.29 ± 37.62 (min 6, max 120) months, 26 received oral steroids (0.5 mg/kg every other day, for 33.04 ± 37.98 (min 1, max 185) months. BMD (g/cm^2) in the lumbar spine (L2–L4) was measured using dual energy osteodensitometry "DPX-MD+", Lunar. All patients received adequate basic therapy including calcium (500–1000 mg/day) and vitamin D (400 IU/day) supplementation.
Results: Mean Z-scores were −1.564, S.D.0.91 (inhaled steroids) and −2.165, S.D.1.75 (oral steroids). Using z-score data, 4/14 (29%) patients inhaled corticosteroids were osteoporotic and 6/14 (43%) were osteopenic. In group of the patients received oral steroids 10/26 (38%) were osteoporotic and 8/26 (31%) were osteopenic. Patients received oral corticosteroids (BMD = 0.635 ± 0.141 g/cm^2) had lower parameters of bone mineral density than children inhalated corticosteroids BMD = 0.839 ± 0.231 g/cm^2) ($p < 0.02$).
We revealed significant correlation between continuance of treatment with oral steroids and BMD ($p < 0.001$). There was no correlation between duration of inhaled steroids therapy and BMD.
Conclusion: Oral corticosteroids therapy is associated with more significant decrease of bone mineral density than use of steroids inhalation. Nevertheless, increased loss of BMD has been expressed in both groups.

350 Audit of renal dysfunction in cystic fibrosis (CF)

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Recent advances in the treatment of CF have led to an increased life expectancy. Conditions which were previously not encountered such as diabetic nephropathy and renovascular disease are likely to become an increasing problem, leading to difficulties in the use of nephrotoxic drugs and complicating transplant suitability.
At our centre clinical incidents involving aminoglycosides led us to look at our prescribing practices and how we monitored renal function in those patients at high risk of nephrotoxicity. Many guidelines exist for the safe prescription of aminoglycosides in CF but there is no consensus as to the best and there are none suggesting how or how often we should review our patients for signs of renal failure.
Over a 2 year period we retrospectively identified 108 patients defined at risk of nephrotoxicity because of CF related diabetes, impaired glucose tolerance or who had $\geqslant 3$ IV antibiotic courses/yr. Prior to this audit, previous prescriptions and drug levels determined dosages. 34 of 108 (31%) had a 24 hr creatinine clearance (Crcl) assessment. Of these 17 (50%) were abnormal with 13 having normal U&Es. Despite this, most had high drug levels at some point in the preceeding year and 3 developed acute renal failure. One patient had hearing loss and high levels were not acted upon in another patient, fortunately without adverse effect.
We cannot rely on historical data for aminoglycoside prescription. Following this audit we have developed a dosing regime based upon 24 hr Crcl (measured or estimated). All Crcl results, drug regimes, dosages and levels are now entered into a database.
Our audit shows the need for regular monitoring of creatinine clearance in patients with CF in order to avoid the excess morbidity and mortality associated with renal disease.